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Posterior Reversible Encephalopathy Syndrome (PRES)

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiographic syndrome that describes certain neuroimaging findings in association with clinical symptoms such as headache, seizure, encephalopathy and vision changes. Classically, PRES is associated with poorly controlled hypertension, and patients present with elevated blood pressure in addition to their symptoms. Most importantly, imaging findings and symptoms are typically reversible, and are a separate entity from ischemic or hemorrhagic cerebrovascular accidents or autoimmune causes of similar symptoms, such as multiple sclerosis [1].

INTRODUCTION

Altered mental status accounts for a large portion of emergency department visits. Its differential is broad and encompasses a multitude of etiologies, from benign to life threatening, and often the cause is multifactorial. As an emergency clinician, it is our duty to differentiate between acute changes in mental state related to neurologic emergency, medical illness, infection, ingestion, psychiatric illness and those related to advanced dementia or delirium [1]. It is often difficult to obtain a reliable history in patients who arrive with altered mental status, and it is essential that a clinician complete a thorough physical exam, medication review and obtain the appropriate labs and imaging when assessing these patients. Occasionally, a thorough history along with physical exam, labs and imaging coalesce to reveal a central cause that explain the patients presenting symptoms.

In this case, we outline a 66-year-old female with no known medical problems who arrived from home via BLS after her friend described “seizure activity.” The history was initially extremely limited, however after a thorough evaluation, the patient was determined to have suffered complications of posterior reversible encephalopathy syndrome (PRES).

CASE PRESENTATION

A 66 year old female with no known medical problems presented to our emergency department via BLS from her home for evaluation of altered mental status following witnessed seizure-like activity by a friend who was visiting her in her home. The episode reportedly lasted “several minutes” and was followed by one episode of emesis and confusion, which persisted on arrival to the ED. Patient was evaluated in the ambulance bay and remained confused, agitated, and speaking in fluent Russian. She did not have any observed focal weakness or deficits. Her vital signs were within normal limits with the exception of a blood pressure of 186/127 and an initial point-of-care glucose was 427mg/dl. During her evaluation, she had a second witnessed tonic-clonic seizure which abated following administration of 2mg IV lorazepam. She was brought immediately to CT scan for imaging to rule out intracranial hemorrhage as a potential etiology of agitation, hypertension and seizure

After her return from CT, the patients daughter arrived at the emergency department and provided further history, and the patient slowly returned to her baseline mental status. Once awake, the patient and her daughter noted that the patient had not seen a doctor in “many years” and has been experiencing intermittent headache, vision changes and “flashing lights” with hallucinations and disorientation multiple times per day over the preceding three weeks.

Physical Examination

Following her return from CT scan, she was re-evaluated and a more thorough examination was performed. General examination revealed a somnolent, obese female who appeared stated age. There was no odor suggesting alcohol intoxication. Her repeat vital signs remained within normal limits, and her blood pressure improved to 155/90 without any intervention. There was dried emesis noted surrounding her mouth, however no intraoral lacerations or evidence of tongue biting was evident. Her sclera were anicteric, pupils were equal, round and reactive to light bilaterally. Her heart, lung, abdominal and evaluation of her extremities was unremarkable. She was somnolent as stated. She withdrew all extremities to painful stimuli, spontaneously moved all extremities and intermittently would purposefully move her extremities as directed.

Results of pathological tests and other investigations

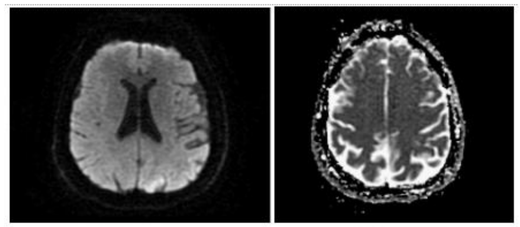
Her labs revealed a white blood cell count of 13.7 and was otherwise unremarkable. Her basic metabolic panel was significant for a glucose of 471 with an anion gap of 25 and a CO2 of 11. Her renal function was within normal limits. Her EKG showed sinus tachycardia at 119 bpm with PVCs. There were no acute ST/T wave abnormalities and her QTC was 475. The remainder of her workup, which included: hepatic function panel, urinalysis, acetone, ammonia, urine drug screen, ethanol, salicylates, acetaminophen and CT head without contrast were all unrevealing.

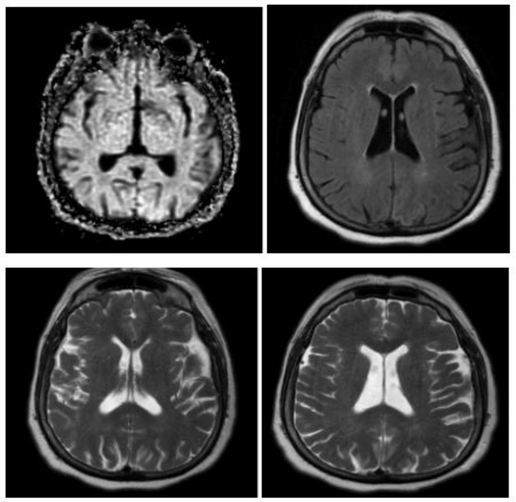
TREATMENT PLAN & OUTCOME

The patient did not require any intervention for her blood pressure during her ED stay, as there was no intracranial hemorrhage and her systolic blood pressure remained at goal between 140-160 to maintain cerebral perfusion. The elevated glucose was thought to be secondary to seizure or stress response, as the patient did not have a known history of diabetes, and she was resuscitated with fluids.

She was admitted to the intermediate care unit for further monitoring, neurology evaluation and EEG monitoring.

The patient was started on anti-hypertensives, cholesterol and diabetes medications. She was scheduled for follow-up with neurology and neuro-ophthalmology as an outpatient. She did not suffer from any recurrent seizures during her hospitalization. As her blood pressure was controlled, she reported less frequent vision changes and her symptoms were resolved prior to her discharge.





MRI interpretation: There is cortical/subcortical edema in the left occipital lobe with some gyral thickening as well as mild increased signal on diffusion images. Ventricles and sulci are otherwise mildly prominent, in keeping with mild cerebral volume loss. There is mild periventricular white matter T2/FLAIR hyperintense signal abnormality, probably related to minimal microangiopathy. No mass effect. No evidence of hemorrhage on gradient echo images. No extra-axial collection. Cerebellar tonsils are normal in location. Major vessel flow voids are preserved in the skull base.
Impression: There is cortical/subcortical edema with gyral thickening as well as increased signal on diffusion images in the left occipital lobe. Etiology may represent subacute infarction, complication of PRES.

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DISCUSSION

Posterior reversible leukoencephalopathy syndrome (PRES) is a hot discussion topic in emergency medicine today. It is most notable for its overall variability and lack of clearly defined clinical features. It was first described in The New England Journal of Medicine in 1996, and has been increasingly diagnosed since - likely due to advances in imaging and physician recognition. Most of the literature that exists on PRES is case based, and clear diagnostic criteria remain elusive.

To better understand this syndrome, we must first understand the proposed pathophysiology. Radiographically and clinically, signs and symptoms of PRES are considered sequelae of white matter vasogenic edema that manifests in the parieto-occipital regions of the brain [3]. Currently, there are two theories regarding how this vasogenic edema is propagated. The first, is that acute elevated arterial pressures cause cerebral hyperperfusion. This temporarily causes dysfunction of the cerebral vascular autoregulatory system, with resultant vascular leakage and vasogenic edema. This is most commonly seen in the posterior circulation, due to the lack of sympathetic tone to the basilar artery. The second proposed mechanism is that extreme hypertension, even transiently, causes vasospasm and local ischemia. This accounts for blood brain barrier endothelial dysfunction and vasogenic or cytotoxic edema on MRI [2,3].

Since PRES cannot be definitively diagnosed without MRI, it is often not diagnosed in the emergency department. Management should include blood pressure control, in accordance with treatment of hypertensive emergencies, with aim of reduction in systolic BP 10-25%. Seizures should be managed with anti-epileptic medications, with benzodiazepines as a first line agent [1]. MRI should be performed as quickly as possible for diagnostic and prognostication purposes.

Emergency clinicians should always keep PRES on the differential for patients presenting with vague neurologic symptoms and elevated blood pressure or poor control of chronic hypertension. A negative CT/CTA does not definitively rule out PRES. Emergency physicians should also keep in mind that while PRES is typically thought to occur with severe hypertension, literature has shown that systolic blood pressure does not always exceed autoregulatory levels (>200 systolic) [3]. As seen in our case, the patient had only transient hypertension that resolved in the emergency department and did not require acute intervention. It was likely the sum of multiple transient elevations in blood pressure that caused her symptoms. The most common presentations in patients with PRES include encephalopathy (28-92%), disorders of consciousness (67-90%), epileptic seizure (70-74%), visual disturbance (20-67%) and headache (26-53%) [2]. Focal neurologic signs are fairly uncommon, and only present in 5-15% of patients with posterior reversible encephalopathy syndrome [2].

CONCLUSIONS

PRES is an important diagnosis in emergency medicine and should be included in all patients with neurologic complaints and elevated blood pressure. Diagnosis of this process requires a high index of suspicion from front line providers given the wide variety of chief complaints that it may encompass. The goal of emergency physicians not to diagnose PRES, but rather to expedite MRI to evaluate for vasogenic edema and assist in the initiation of long term medical management to prevent permanent neurologic injury.